Eur J Vasc Endovasc Surg (2011) 42, 381–383

SHORT REPORT

Diffuse Dermal Angiomatosis

K. Morimoto [a,*], H. Iioka [a], H. Asada [b], K. Kichikawa [c], S. Taniguchi [d], M. Kuwahara [a]

[a] Department of Plastic Surgery, Nara Medical University, 840 Shijo, Kashihara, Nara 634-8521, Japan
[b] Department of Dermatology, Nara Medical University, 840 Shijo, Kashihara, Nara 634-8521, Japan
[c] Department of Radiology, Nara Medical University, 840 Shijo, Kashihara, Nara 634-8521, Japan
[d] Department of Cardiovascular Surgery, Nara Medical University, 840 Shijo, Kashihara, Nara 634-8521, Japan

Submitted 19 February 2011; accepted 11 May 2011
Available online 8 June 2011

KEYWORDS
Diffuse dermal angiomatosis;
Atherosclerosis;
Revascularization

Abstract Diffuse dermal angiomatosis (DDA) is characterized clinically by painful erythematous lesions with ulcers and histologically by a benign, diffuse, and self-limited proliferation of tiny blood vessels in the superficial layers of the reticular dermis. Here we describe a case of DDA with leg ulcer. Erythematous lesions presented around the ulcer and angiogram revealed an occlusion of the superficial femoral artery. The erythematous lesions disappeared after revascularization. Although DDA is extremely rare, early correction of the ischemia in the peripheral artery should be taken into consideration.

Introduction

Diffuse dermal angiomatosis (DDA) is a rare, acquired cutaneous, reactive, vascular disorder that was first described in 1994.[1] DDA is a benign vascular proliferation characterized by poorly circumscribed, erythematous to violaceous lesions with frequent ulceration. Histologically, diffuse and mostly extravascular interstitial proliferation of CD31-positive endothelial cells and myofibroblasts are present within the reticular dermis in DDA cases.

* Corresponding author. Tel.: +81 744 29 8890; fax: +81 744 25 8511.
E-mail address: keisukemorimoto2@live.jp (K. Morimoto).

Case Report

A 65-year-old male with a history of hypertension and smoking (smoking a pack per day for 40 years) presented with a 1-month history of a painful, erythematous lesion on the right lower extremity (Fig. 1A,B). Over the past 6 months, he noticed progressive intermittent claudication of the right calf after walking approximately 300 m. However, the walking distance without pain had gradually shortened, and decreased to 50 m at the first admission. He had non-palpable pulses in the right popliteal, posterior tibial, and dorsal pedal positions. In addition, the right ankle-brachial index was unmeasurable and the right toe systolic pressure was 20 mmHg.

After 1 week, he developed an ulceration on the lesion. Prednisolone (10 mg/day) was administrated, as is

1078-5884/$36
doi:10.1016/j.ejvs.2011.05.011

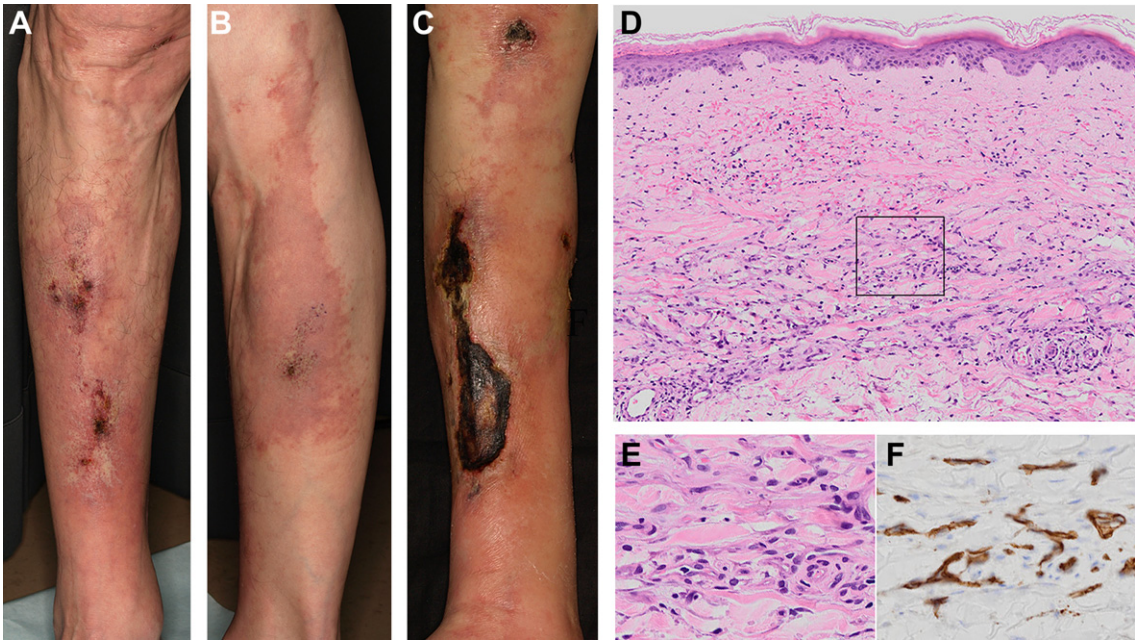

Figure 1 A: Erythematous lesion on the front right leg with purpuric lesion. B: The back of the right leg. C: Leg ulceration progressed. D, E: Histopathologic examination demonstrates a diffuse proliferation of benign endothelial cells in the papillary and reticular dermis. There is scant formation of small vascular lumina with occasional extravasating erythrocytes and no evidence of cholesterol emboli or vasculitis (A and B, hematoxylin—eosin stain; original magnification: A, 40×; B, 400×). F: Positive immunohistochemical staining using anti-CD31 antibody confirms the endothelial nature of the infiltrate (original magnification 400×).

commonly prescribed in patients with vasculitis. A blood test was performed, and a biopsy was obtained from the central, non-ulcerated region of the lesion. Values of antinuclear body, rheumatoid factor, lupus anticoagulant, anticardiolipin body, cryoglobulin, cytoplasmic anti-neutrophil antibody (C-ANCA), cytoplasmic perinuclear anti-neutrophil cytoplasmic antibodies (P-ANCA), fibrin degradation products (FDP), and D-dimer were normal. Histopathologic examination demonstrated a dense proliferation of endothelial cells (Fig. 1D—F). Staining was negative for human herpes virus 8. The clinical and pathological findings were consistent with DDA, and therefore

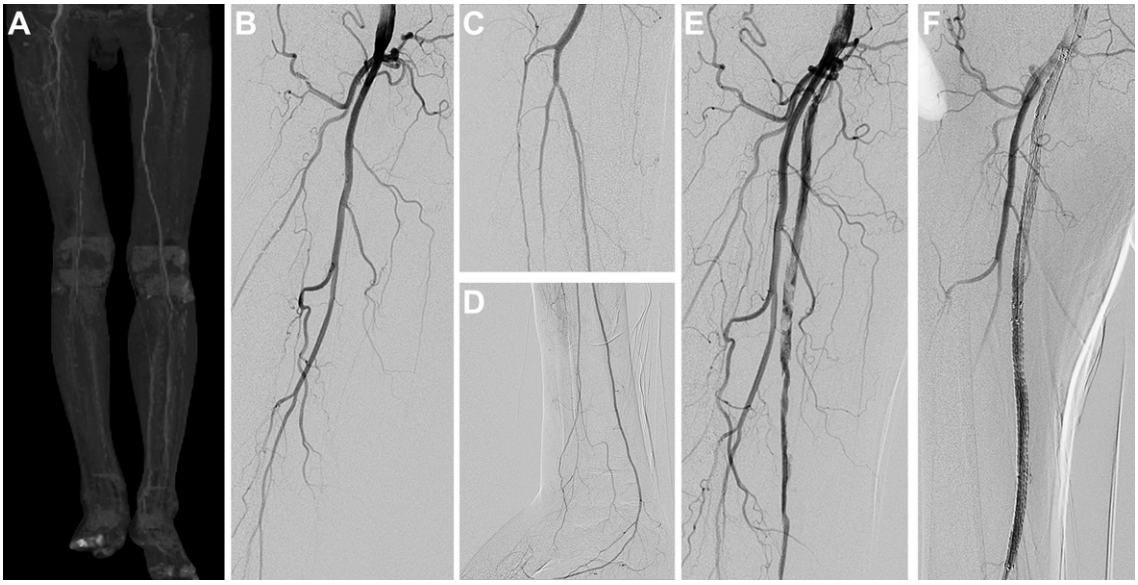

Figure 2 A: CT angiography showed the right superficial femoral artery occlusion and collateral blood flow. B: Angiogram revealed an occlusion of the right superficial femoral artery. C, D: Distal run off was well seen despite partial stenosis at anterior tibial artery. E: Several stenotic lesions were noted after thrombolysis. F: Endovascular stent was deployed distal to the superficial femoral artery.

prednisolone was reduced and stopped. A subsequent CT angiography showed right superficial femoral artery occlusion and collateral blood flow (Fig. 2A). Doppler ultrasonography showed the presence of thrombosis at the superficial femoral artery.

The patient's ulcer continued to grow (Fig. 1C) and he was admitted 4 weeks after presentation. DSA before the procedure showed a 20-cm-long occlusion of the right superficial femoral artery from the orifice (Fig. 2B—D). He was diagnosed with acute thrombosis, and 12×10^4 units of urokinase were infused from the catheter and recanalization was obtained. Residual thrombi were aspirated using the Thrombaster III (Kaneka Medix, Osaka, Japan). However, several stenotic lesions were noted (Fig. 2E), and therefore balloon angioplasty and implantation of a 6×120 mm Luminexx stent (Bard Peripheral, Vascular, Tempe, AZ) were performed (Fig. 2F). The right ankle-brachial index increased to 1.01 and toe systolic pressure to 120 mmHg. Following the revascularization, the patient was prescribed oral aspirin (100 mg/day) and sarpogrelate hydrochloride (300 mg/day). Five weeks after the revascularization procedure, the erythematous lesions on the right lower extremity disappeared and the skin ulcer region was reduced.

Discussion

To date, 7 cases of DDA with atherosclerosis including our case have been reported,[1—5] with similar clinical and histopathological findings. All patients presented with erythematous or violaceous lesion with ulcer and had arterial stenosis. All underwent a revascularization procedure, which resulted in the healing of the erythematous lesion and skin ulcer region within 10 weeks.

The 6-month-old intermittent claudication and the presence of collateral circulation with several stenotic lesions on the angiogram suggested that leg ischemia was caused by chronic arterial disease. Moreover, if proliferation of endothelial cells resulted in chronic ischemia, the patient would demonstrate necrosis, but not proliferation of endothelial cells, with acute ischemia. The age of the thrombus in the superficial femoral artery is unknown, but based on the fact that lysis was successful, it is unlikely to be older than 2—3 weeks. It is reasonable to assume that stenosis was present prior to the acute event, and acute widespread thrombosis at the superficial femoral artery might have the potential to attenuate the rapid progression of the ulcer. Thus, DDA might be associated with severe ischemia, because all published cases of DDA-related atherosclerosis had severe ischemia with ulcerations. Furthermore, we found that the emergence of DDA was synchronized with the deterioration of ischemia. DDA might presage the emergence of ischemic ulcer.

In summary, we described a case of DDA-related severe peripheral vascular atherosclerosis. Successful revascularization facilitated the healing of the DDA in our patient, as well as in all published cases. DDA might be a premonitory sign of ischemic ulcer. When DDA is found in patients with peripheral vascular disease, early correction of the ischemia should be taken into the consideration.

Acknowledgments

We thank Dr D. Tsuruta, Department of Dermatology, Osaka City University of Medicine, for many helpful suggestions.

Funding

N/A.

Conflict of Interest/Funding

None.

References

1 Krell JM, Sanchez RL, Solomon AR. Diffuse dermal angiomatosis: a variant of reactive cutaneous angioendotheliomatosis. *J Cutan Pathol* 1994;**21**:363—70.
2 Kimyai-Asadi A, Nousari HC, Ketabchi N, Henneberry JM, Costarangos C. Diffuse dermal angiomatosis: avariant of reactive angioendotheliomatosis associated with atherosclerosis. *J Am Acad Dermatol* 1999;**40**:257—9.
3 Kim S, Elenitsas R, James WD. Diffuse dermal angiomatosis: a variant of reactive angioendotheliomatosis associated with peripheral vascular atherosclerosis. *Arch Dermatol* 2002;**138**:456—8.
4 Draper BK, Boyd AS. Diffuse dermal angiomatosis. *J Cutan Pathol* 2006;**33**:646—8.
5 Kirkland CR, Hawayek LH, Mutasim DF. Atherosclerosis-induced diffuse dermal angiomatosis with fatal outcome. *Arch Dermatol* 2010;**146**:684—5.